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Multidimensional Analysis Of Advanced-Stage Huntington's Disease From Neurocognitive And Psychofunctional Perspectives With Morphometric Correlations: Case Series

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Introduction

- Huntington's disease (HD) is a progressive neurodegenerative monogenic disorder, and its multifaceted clinical and radiological analysis correlations are not yet understood.
- We aim to evaluate advanced-stage HD patients for multidimensional clinical deterioration with objective scales and correlate this with morphometric-based measurements

Materials and Methods:

- Ten advanced-stage HD patients evaluated with the Unified Huntington's Disease Rating Scale (UHDRS) were subjected to psychofunctional assessment for behavioral and neurocognitive decline, total functional capacity (TFC), and functional assessment scale (FAS) for functional determination.
- In the morphometric assessment, bicaudate ratio (BCR), bi-frontal ratio (BFR), frontal horn area (FHA), frontal horn ratio to intercaudate distance (FH/CC), and caudate volume and caudate volume ratio (CVR) were analyzed and correlated with relevant parameters (Figure).

Results

- The most frequent functional decline was observed for occupational and financial ability in UHDRS TFC (5.60±2.27), social/financial engagement, and self-care impairment in the FAS (11.10±3.48).
- Cognitive decline was especially prevalent in quick thinking and responding to stimuli on time and to a sufficient extent. Caudate volume loss was more severe on the right-hand side (6.50±1.18) and inferior sections(21.65±7.30).
- A negative correlation was found between intercaudate distance and the verbal fluency test (rho=-0.775).
- The Parkinson's disease sleep scale (PDSS) and intercaudate distance were negatively correlated (rho=-0.559), and a positive correlation was found for the bi-frontal distance/caudate distance (rho=0.559). There was a negative correlation between the Questionnaire for Impulsive–Compulsive Disorders in Parkinson's Disease-Rating Scale (QUIP-RS), the Hamilton Depression Rating Scale (HDRS), and the Hamilton Anxiety Rating Scale (HAM-A) and frontal horn distance (FHD) (rho=-0.671, rho=0.61 and rho=0.571, respectively). (Table)

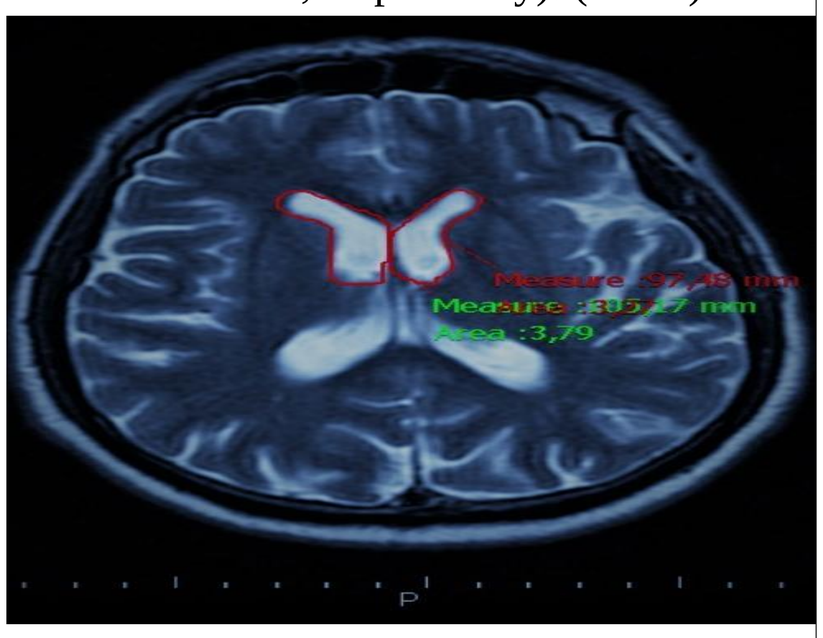

Figure : Morphometric assessment of FHA.

Discussion

- HD is an autosomal dominant neurodegenerative disorder caused by an increasing number of cytosine-adenine-guanine (CAG) repeats in the huntingtin gene (HTT).
- Motor dysfunction (1) and cognitive decline (2) can be effected early in the HD which is compatible with regional brain volumetric losses.
- Also neuropsychiatric alterations have been reported in correlation with a heterogeneous episodic pattern (3).

Table . Correlation analyses of neuropsychiatric findings and MRI

		Neuropsychiatric Scales				
		PDSS	QUIP-RS	SAS	HDRS	HAM-A
Magnetic Resonance Imaging Morphometric Evalution	Intercaudate Distance (mm)	rho=-0,559 p=0,043	rho=-0,345 p=0,329	rho=0,477 p=0,164	rho=0,274 p=0,443	rho=0,286 p=0,424
	Frontal Horn Distance (mm)	rho=-0,073 p=0,841	rho=-0,671 p=0,034	rho=0,483 p=0,157	rho=0,610 rho=0,571 p=0,021	rho=0,571 p=0,034
	Bifrontal Distance/ Caudate Dsitance	rho=0,559 p=0,043	rho=0,098 p=0,787	rho=-0,194 p=0,590	rho=-0,256 p=0,475	rho=-0,255 p=0,476
	Frontal Horn Area (Right, mm)	rho=-0,152 p=0,675	rho=-0,56 rho=-0,622 p=0,042	rho=0,401 p=0,250	rho=0,482 p=0,159	rho=0,486 p=0,154
	Frontal Horn Area Left , mm)	rho=-0,152 p=0,675	rho=-0,622 p=0,035	rho=0,552 p=0,048	rho=0,268 p=0,454	rho=0,316 p=0,374

Conclusion

- In light of the current findings, caudate atrophy is an important indicator of cognito-functional disability, especially in terms of verbal ability.
- The right hemisphere seems to be more vulnerable to neurodegenerative processes, and mood disorders appear to be related explicitly to right frontal lobe degeneration.
- Psychofunctional deterioration may begin years before clinical diagnosis, so HD should be considered in the differential diagnosis of aberrant psychofunctional deterioration in young patients..

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